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CASE REPORT

Behçet's Aortitis – a Case Report: False Aneurysm Rupture and Aortico-duodenal Fistula

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Introduction

A multisystemic inflammatory chronic disorder, Behçet's disease manifested as a triform of relapsing iritis stomatitis and urogenital ulceration, and is now recognised as a systemic vasculitis that affects both veins and arteries.[1] Originally described in 1937 by Hulusi Behçet,[2] this disease has its highest incidence in Japanese and Eastern Mediterranean populations. It appears often in the third or fourth decade of life.[1–4] It is generally accepted that surgical results of inflammatory arterial aneurysms are not as favourable as the surgical treatment of the atherosclerotic aneurysms. This situation more strikingly exists for Behçet's disease.[4]

Many complications have been recognised for Behçet's disease. In this report, we present the case of a young man with Behçet's aortitis, one of the most severe manifestations.

Case Report

A 38-year-old Caucasian male with back pain for 1 month visited our outpatient clinic. He had the diagnosis of Behçet's disease more than 10 years previously and was hospitalised because of chronic pancreatitis 1 year ago in another centre. He was treated for iritis 10 years ago, and for genital ulcerations 5 years ago, experiencing relapses. Computerised tomography was performed for differential diagnosis on 21 March 1997, when he was hospitalised for chronic pancreatitis. Abdominal aorta and vascular structures seemed to be normal at that time. One year after his discharge he revisited our clinic with abdominal pulsatile and sensitive mass.

On 24 March 1998 abdominal ultrasonographic evaluation was performed, revealing an infrarenal abdominal pseudoaneurysm. The following day computerised tomography confirmed the diagnosis and showed the deplacement of the aorta to left anterior, right renal artery to the anterior and compression to vena cava distally. There was a site rupture at posterior location also. The dimensions were 7 cm in length and 5 cm in diameter. Digital substraction angiography (DSA) was performed, diagnosis was confirmed and renal artery anomaly detected.

On 26 March 1998 the patient was operated on as an emergency. The midline incision was performed. There were para-aortic lymphadenopathies and dense inflammations. The pseudoaneurysm was ruptured posteriorly. Infrarenal pseudoaneurysm was resected and replaced with 20 mm collagen sealed tube graft (Hemashield, Meadox). Left accessory renal artery was reimplanted to the graft. After haemostasis the aneurysm was oversewn primarily. No complications occurred.

The patient spent four days in an intensive care unit and began oral intake on the third postoperative day. He was discharged with uneventful recovery on the seventh postoperative day. The histopathological definition of the intraoperative aortic tissue biopsy was diagnosed as degenerative necrotising vasculitis. There was intimal irregularity at the vascular wall, endothelial proliferation, lymphocyte and polymorphonuclear leucocyte infiltration.

In October 1998 the patient was re-admitted to hospital because of fever, lumbago and bilateral lower

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extremity pain. In January 1999, the patient attended hospital with an abscess formation at the left popliteal region, proximal site of gastrocnemius muscle. The results of the cultures from the abscess and blood revealed enterococci. After the drainage of the abscess and proper antibiotic therapy the symptoms disappeared and the abscess healed. DSA control was performed showing no problem via abdominal graft.

A year after the first operation, the patient visited our out-patient clinic with haematemesis and melana that was continued in the previous week. His haematocrit was 26% and 40 units of bank blood were given. Endoscopic evaluation was performed and aorta duodenal fistula was diagnosed. There was invasion of the aortic graft into the duodenum. Second operation was performed on 10 June 1999. Duodenum was repaired primarily and the infected graft (via enterroccocci) was resected and replaced with a Rifampicine-bound polyester tube graft (intervascular). Also, omentoplasty was performed. On 16 September 1999, 3 months after the second operation, the patient suffered abdominal pain. Abdominal ultrasonography showed 9×8 cm pseudoaneurysm on the proximal and distal end of the graft. DSA demonstrated the pseudoaneurysm at the proximal anastomosis site of the graft. While waiting in the ward for the consensus, the patient's abdominal pain worsened, with rebound sensitivity and rupture of the false aneurysm. He was operated on in a semi-urgent manner and a descending aorta biiliac "Y graft" was interposed. Proximal anastomosis was made to aorta over the diaphragm. Distal anastomosis was performed at the common iliac arteries, infrarenal exgraft was excised and aortic remnant tissue was debrided.

In January 2000, the patient visited our out-patient clinic for the first control. Neither physical nor ultrasonographic and angiographic evaluation gave any pathological result.

Comment

The incidence for cardiovascular manifestations of Behçet's disease is approximately 30% and appears to be the major cause of death. The underlying pathological process was thought to be a vasculitis affecting the vasa vasorum, resulting from fragmentation and splitting of elastic fibres in the media layer with perivascular mononuclear cell infiltration.[3]

It is generally accepted that surgical results of inflammatory arterial aneurysms are not as favourable as the surgical treatment of the atherosclerotic aneurysms. This situation more strikingly exists for Behçet's disease.[4] Statistically the major causes of death in Behçet's disease are cerebrovascular accidents, ischaemic bowel perforations and aneurysm ruptures which are mainly arterial complications.[4] Actually, even though it is a systemic inflammatory vasculitis, there isn't any evidence that it appears as panaortitis. Usually local ulcerations and perforation at the ulcer sites cause the dramatic manifestations. As is the case in the patient presented here, aneurysms are generally false in nature. Contained rupture of the false aneurysm is usually the manifestation of the disease.

Few reports have addressed the use and effectivity of steroid therapy for control of any inflammatory disease with critical side-effects such as susceptibility to infection and delayed wound healing.[6] It is important to cause the inflammatory reaction to subside pre- and postoperatively, to reinforce the suture line and implanted prosthesis, and to select the appropriate operative procedure, bypass conduits in cardiovascular disorders.

The abdominal aorta is one of the sites affected most often, and its reconstruction is often followed by haematemesis or melena due to graft-enteric fistula by anastomic pseudoaneurysmal formation and its rupture.[7] In Behçet's disease, even the angiographically smaller aneurysms may enlarge rapidly to rupture, resulting in a quick fatal bleeding during the period of best medical treatments. In addition, a pseudoaneurysm often occurs at the anastomotic site of an artificial graft.[7] Therefore, patients with an aneurysm of Behçet's disease must be followed intensively and must be treated surgically without overlooking any clinical evidence of acute pseudoaneurysmal expansion or an aneurysmal rupture.[1]

Aorta-enteric fistula is an uncommon but fatal complication of vascular surgery, manifesting with upper gastrointestinal bleeding, sometimes massive, threatening the patient's life, occuring within months to years after the original operation, with an incidence of 0.4–4%.[8]

Homografts and synthetic arterial substitutes have shown degenerative properties. Erosion should be prevented at the initial operation by avoidance of contamination by meticulous suture technique and by interpositioning of viable tissues (omentum, retroperitoneal fat, peritoneum, aneurysm wall) between the graft and the adjacent bowel.[8] Secondary aorto-enteric communication should be suspected when upper gastrointestinal bleeding occurs after aortic surgery. The parts of the alimentary tract usually involved are the third and fourth portions of the duodenum (60%), the ileum in the 18%, the caecum in 12%, the sigmoid in 8% and the remaining gastrointestinal tract

in 2%.[8,9] Upper gastrointestinal tract endoscopy seems to be of help since it may reveal the fistula or rule out other causes of intestinal bleeding. In our patient, we have seen the graft in duodenum with endoscopy.[8] Total removal of the graft is mandatory if on the one hand infection is considered infected and on the other the intestine might have adhered to the graft. The duodenal or jejunal defect is usually closed transversely with standard intestinal suture techniques.

Since the results of surgery have lots of pitfalls in Behçet's disease, interventional semi-invasive procedures may be more reasonable in these patients. Endovascular techniques may have more favourable results than surgical procedures to avoid post-surgery complications.

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